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ORIGINAL ARTICLE

Imaging of Cardiac Tumors and Masses

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KEY WORDS: *Cardiac tumors, cross sectional imaging modalities, computed tomography, magnetic resonance imaging, positron emission tomography*

ABBREVIATIONS
CT = computed tomography
MRI = magnetic resonance imaging
PET/CT = positron emission tomography

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ABSTRACT

OBJECTIVE: As noninvasive imaging is gaining widespread acceptance, the aim of the present study was to review our experience with the use of cross sectional imaging modalities to evaluate cardiac tumors and masses.

BACKGROUND: Proper diagnosis of the type and extension of a cardiac tumor or mass is very important for therapy planning. Echocardiography has an established role as a non-invasive diagnostic imaging modality. Computed tomography (CT) and magnetic resonance imaging (MRI) also provide adequate delineation of cardiac tumors and masses.

METHODS: Twenty-two patients referred to our department with a suspected cardiac mass, previously detected by cardiac ultrasound, were evaluated by CT (12 patients) and MRI (4 patients). Six patients were examined by both modalities. In 2 cases of staging of a known primary neoplasm and one case of a suspected epicardiac lesion, positron emission tomography (PET-CT) was performed. The CT cardiac examinations were performed using a 16-slice multislice scanner with ECG gating. The MRI examination was performed on a 1.5 Tesla MRI scanner using an ECG-gated cardiac protocol and the PET-CT examination was performed on an integrated PET-CT four-slice scanner, using 370 mMBq of fluorodeoxyglucose.

RESULTS: Twenty-two cardiac masses were successfully detected and their characteristics adequately delineated, including eight myxomas, one angiosarcoma, one ventricular lymphoma, one endocardiac metastasis, one epicardiac paraganglioma, and 10 cases of intracardiac thrombi.

CONCLUSION: CT and MRI are noninvasive imaging modalities which can delineate cardiac tumors and masses and provide essential information for adequate diagnosis, staging and treatment planning. Compared to cardiac ultrasound, CT and MRI are superior in preoperative planning.

INTRODUCTION

Cardiac tumors are rare in everyday clinical practice; nevertheless, they represent an important group of tumors that require investigation for proper diagnosis and treatment. Echocardiography has been the only reliable non-invasive diagnostic tool

until the past decade. Although computed tomography (CT), magnetic resonance imaging (MRI) and positron emission tomography (PET-CT) are gaining widespread acceptance, CT can locate, differentiate, confirm or exclude the presence of a cardiac mass and it is thus regarded essential for adequate staging.[1] Additionally, it provides detailed information of cardiac and vascular anatomy related to the mass, which allows proper therapy planning. The role of MRI has also been demonstrated in the assessment and differential diagnosis of cardiac tumors, inflammatory disease, storage disease and cardiomyopathies.[2] Both modalities may confirm or exclude the presence of intra-cardiac thrombi. Additionally cardiac function and flow may be evaluated by MRI.

The aim of the present study was to review our experience with the use of these newer imaging modalities in evaluating cardiac tumors and masses in 22 patients referred for further evaluation after an initial echocardiographic examination.

METHODS

We examined 22 patients (17 males and 5 females), referred to our department from April 2004 until November 2009, with a suspected cardiac mass detected by cardiac ultrasound. The review of the records of these patients, as well as the publication of the data of this study was approved by the hospital ethics committee. A CT scan was performed in 12 patients, MRI in 4 patients and 6 patients were examined by both modalities. A PET-CT scan was performed in one patient with a suspected epicardiac lesion detected by echocardiography and another patient for staging of melanoma.

All CT cardiac examinations were performed using a 16-slice multislice scanner, with ECG gating. Reconstructions of images were performed between phase 70% and 75% of the cardiac cycle due to better spatial resolution. Image post-processing was performed on a workstation. Images were acquired before the injection of contrast medium in order to evaluate calcifications and were followed by the injection of non-ionic iodinated contrast medium, 370 mgI/ml, using an injector with a rate of 3.3-3.5 ml/sec through an antecubital vein; 20 ml of saline solution was injected to flush the vein (saline chaser).

The MRI examinations were performed on a 1.5 Tesla MRI scanner using an ECG-gated cardiac protocol. The examination started with a morphological overview, which covered the entire heart by using fast spin-echo sequences, single-shot gradient-echo sequences and steady-state free precession sequences, reducing the examination time. Initial morphological analysis was followed by soft tissue characterization techniques, T1-weighted pre- and post contrast medium injection and T2-weighted fast spin-echo sequence. These techniques were applied to the region of the tumors.[1] T1-weighted, T2-weighted, inversion recovery sequences and T2 gradient-echo sequences were also used for identifying the presence of possible calcifications or hemorrhages in the tumor. Enhancement evaluation of the tumors was performed by injection of contrast medium 0.1 mmol/kg by an injector through an antecubital vein. Delayed- enhancement gradient-echo sequences were performed to define the extent of the mass. Post-processing was performed on a workstation.

The PET-CT examination was performed on an integrated PET-CT four-slice scanner. A total of 370 mMBq of fluorodeoxyglucose (FDG) was injected intravenously one hour before the examination was initiated and all PET and CT images were attenuation corrected and fussed in three planes and were evaluated on a workstation.

RESULTS

Twelve cardiac tumors were detected including eight myxomas (Fig. 1-4), one angiosarcoma (Fig. 5), one ventricular lymphoma (Fig. 6), one endocardiac metastasis (Fig. 10) and one epicardiac paraganglioma (Fig. 7). Ten cases of intracardiac thrombi were detected (Fig. 8 & 9). The lymphoma case was further proven by biopsy; the myxomas, sarcoma, and paraganglioma were surgically confirmed. Three more metastases were detected by PET-CT in the patient with the melanoma metastases.

The exact location of the tumors and masses detected was as follows: eight myxomas in the left atrium, four thrombi in the left ventricle and six in the right atrium. The angiosarcoma was located in the right atrium. One lymphoma was detected in the right ventricle, the paraganglioma on the epicardium, specifically in the left atrioventricular groove and a metastasis was detected in the endocardium of the left atrium. The CT and MRI findings were similar with those of echocardiography with regards to the myxomas and thrombi, while small differences concerning mass sizes were reported.

The size of the myxomas ranged between 1.5 and 6 cm. Three of them were partially calcified and one was totally calcified (Fig. 4), two were papillary and two were prolapsing through the mitral valve. The size of the intracardiac thrombi ranged between 1 and 4 cm. Seven thrombi were located in the right atrium (one particularly in the crista terminalis) and the remainder (n=3) in the left ventricle. A left ventricular aneurysm was detected in two of these cases post myocardial infarction. The angiosarcoma was located in the right atrium and infiltrated the atrial septum and the pericardium causing hemopericardium (Tables 1 & 2).

DISCUSSION

Echocardiography fulfils the clinician's expectations in the vast majority of cases in the detection of cardiac masses.

FIGURE 1A, B. CT: Left atrium myxoma prolapsing in the left ventricle, through the mitral valve.

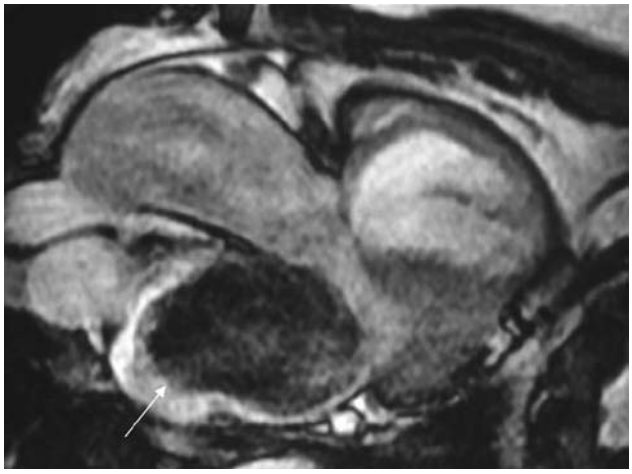

FIGURE 2. MRI: Left atrium myxoma.

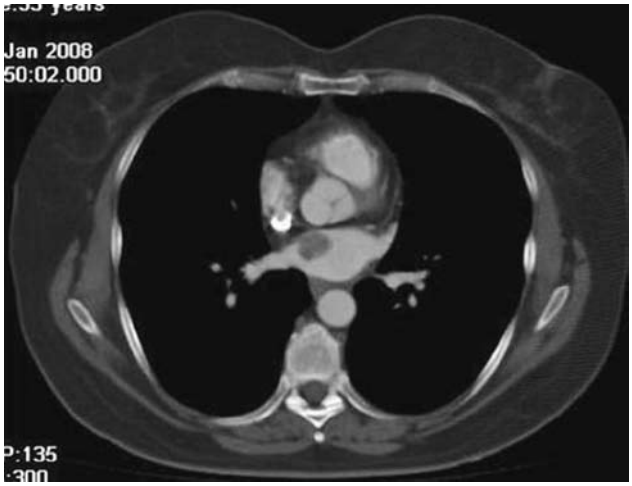

FIGURE 3. Small pendunculated myxoma in the left atrium.

Availability, cost, operator expertise and the image quality of ultrasound equipment today are all favoring echocardiography and this method remains the key examination in assessing cardiac tumors. However, echocardiography is still dependent on the patient's acoustic window.

The advantage of MRI compared to CT is that it does not use ionizing radiation and can evaluate the function of the heart more reliably, while compared to echocardiography that it is independent of anatomical windows. The MRI techniques can produce 3D reconstructions of the heart oriented in any plane and with high contrast between muscular structures and blood.[3,4] MRI provides a complete evaluation of the lesions involving cardiac chambers and the pericardium providing diagnostic information useful for surgical planning. Details of the tumors assessed are morphology, dimensions, location, extension, presence of infiltration, and any signs that may allow characterization, such as adipose tissue, necrosis, hemorrhage, vascularization, calcifications, and myxoid tissue. Most often, diagnosis is based upon indirect and probabilistic findings, which can point to a specific type of tumor rather than another. In general, MRI is superior to CT due to better tissue characterization and lack of ionizing radiation, a very important factor, especially when applying imaging techniques to children. Functional information concerning wall motility, valve function and flow may also provide information on tumors.

Echocardiography, scintigraphic methods and digital subtraction angiography are established modalities in the diagnosis of cardiac masses. With the introduction of multislice CT with ECG-gating, temporal and spatial resolution of CT has improved. Computed tomography also identifies unexpected cardiac tumors in patients examined for other

FIGURE 4. Calcified myxoma in the left atrium protruding in the left ventricle.

FIGURE 5. Right atrium angiosarcoma infiltrating the atrial septum and the pericardium causing hemopericardium.

thoracic pathology.

Cardiac tumors are classified as primary or secondary neoplasms due to metastasis or extension of malignancies located outside the heart. The prevalence of primary tumors is 0.0017-0.33%, whereas secondary tumors occur 20-40 times more often.[1,5]

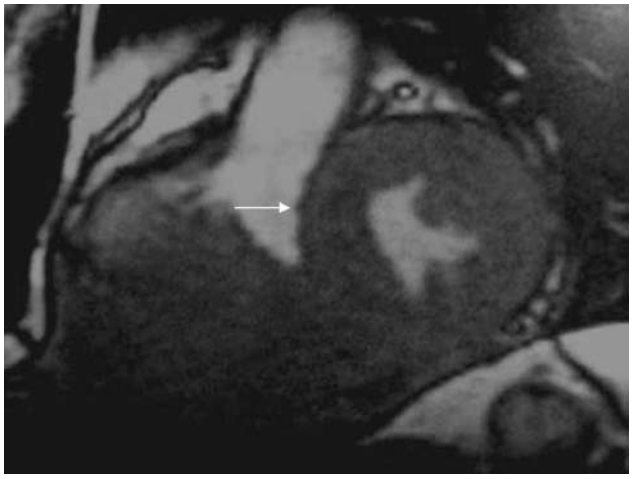

FIGURE 6. MRI: Lymphoma of the left ventricle infiltrating the myocardium.

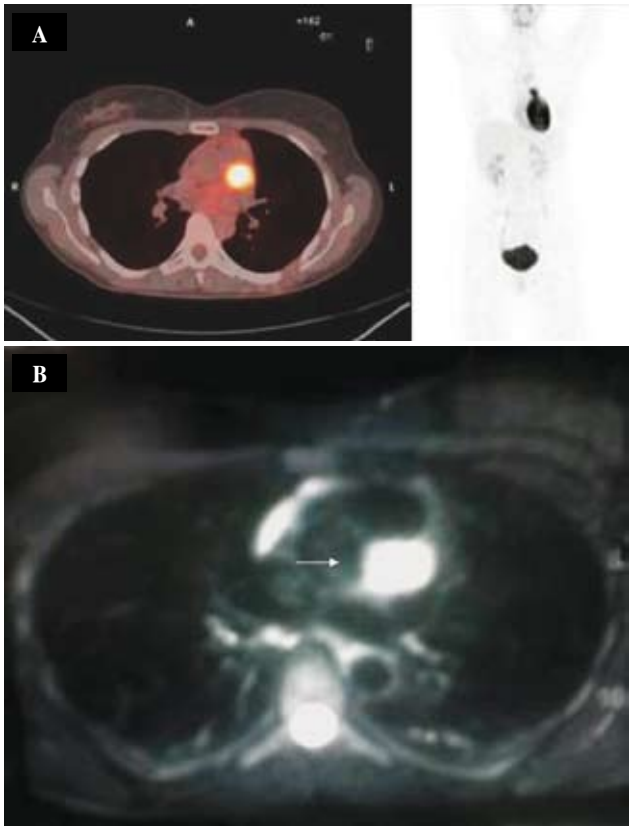

FIGURE 7. A. PET/CT: Paraganglioma detected on the epicardium, atrioventricular (AV) groove. **B.** MRI: High intensity lesion detected in the AV groove.

Neoplastic disease of the heart, whether primary or secondary, can present with symptoms and signs related to involvement of the pericardium, myocardium, and the cardiac cavity leading

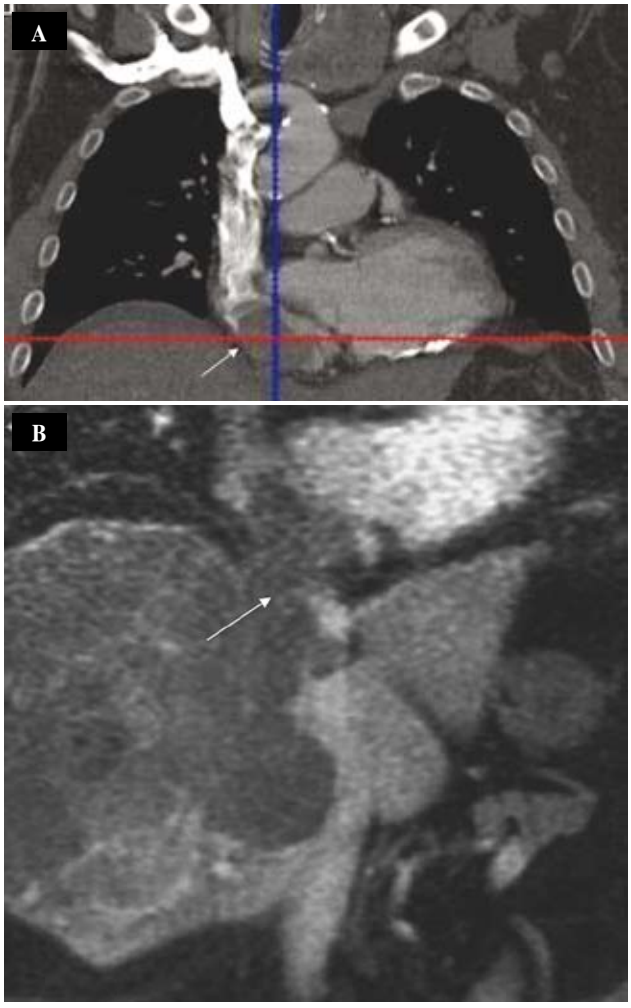

FIGURE 8 A, B. Massive venous thrombosis with cardiac invasion as primary manifestation of hepatocellular carcinoma. Coronal view CT (a), MRI (b).

FIGURE 9. CT: Thrombus in the right atrium (crista terminalis).

FIGURE 10. PET/CT: Metastasis in the left atrium from melanoma.

to cavity or valve obliteration, but can also produce embolic phenomena. With pericardial involvement, the following can be observed: pericarditis, hemopericardium, tamponade as in our angiosarcoma case (Table 1), and arrhythmias. Myocardial involvement can present with arrhythmias, congestive heart failure as in our lymphoma case, angina and infarction due to coronary involvement.[2] Intracavitary masses may mimic mitral stenosis; our two cases of prolapsing myxomas presented this way.

The most common primary cardiac neoplasm is the benign *myxoma*. It accounts for 30-35% of all primary cardiac tumors and for approximately 50% of all benign primary cardiac masses. The typical age of involvement is between 30 and 60 years of age. Clinical presentation may mimic that of mitral valve disease. Hemolytic anemia is typically associated with

TABLE 1. Clinical and imaging characteristic of the tumors

	Gender	Age	Diagnosis	Modalities used	Modality of confirmation	Characteristics of the lesions
1	Male	58	Myxoma	US, CT	CT	Totally calcified, prolapsing, 6 cm
2	Male	71	Myxoma	US, CT	CT	Partially calcified, 3.3 cm
3	Male	65	Myxoma	US, CT	CT	Partially calcified, 2.7 cm
4	Male	63	Myxoma	US, CT	CT	Homogenous, prolapsing, 5 cm
5	Female	55	Myxoma	US, CT	CT	Pendunculated, 2 cm
6	Female	59	Myxoma	US	MRI	Pendunculated, 1.8 cm
7	Male	63	Myxoma	US, CT, MRI	MRI	Partially calcified, 1.5 cm
8	Male	57	Myxoma	US, CT	MRI	Homogenous, solid, 3.5 cm
9	Male	73	Thrombus	US, CT	CT	Left ventricle, 2.5 cm
10	Male	72	Thrombus	US, MRI	MRI	Right atrium, 3.4 cm
11	Male	66	Thrombus	US, MRI	MRI	Right atrium, 1 cm
12	Male	69	Thrombus	US, MRI	MRI	Left ventricle, 4 cm
13	Female	52	Thrombus	US, CT, MRI	MRI	Right atrium, 2.9 cm
14	Female	49	Thrombus	US, CT	CT	Crista terminalis, 1.5 cm
15	Female	61	Thrombus	US, CT	CT	Left ventricle 1.8 cm
16	Male	62	Thrombus	US, CT	CT	Left ventricle, 2.7 cm
17	Male	58	Thrombus	US, MRI	MRI	Right atrium, 3.2 cm
18	Male	66	Thrombus	US,CT, MRI	MRI	Right atrium
19	Male	67	Angiosarcoma	US	CT	Infiltration of atrial septum and pericardium causing hemopericardium
20	Male	62	Lymphoma	US, PET-CT	MRI	Left ventricle infiltration
21	Male	59	Metastasis	US, PET-CT	PET-CT	Left atrium, 1 cm
22	Male	55	Paraganglioma	US	PET-CT	Epicardium, atrioventricular groove, 1.3 cm

CT= computed tomography; MRI= magnetic resonance imaging; PET-CT= positron emission tomography; US= (cardiac) ultrasound

calcified myxomas. Atypical symptoms include syncope, weight loss, fatigue, fever, anemia, an elevated sedimentation rate and the presence of abnormal serum proteins.[2] A myxoma syndrome is also described which is characterized by skin pigmentation and the presence of endocrine neoplasms. Tumor embolism occurs in approximately 50% of cardiac myxomas.[6]

Most of the myxomas (95%) are located within the atria, at the interartial septum at the level of the fossa ovalis, most commonly are left sided (75%), followed by the right atrium (20%) and the remainder may be encountered in either ventricle. They are typically attached to the oval fossa by a stalk. A prolapsing myxoma may impair ventricular filling. They range in size from 1 to 15cm (average 5 cm) and may contain blood, thrombus or calcifications.[2,7] The location, appearance and mobility of myxomas are characteristic. However, large vegetations, thrombus and other tumors must be excluded. Imaging modalities such as CT and MRI can provide additional information when echocardiography cannot delineate the extent of cardiac wall involvement. If the quantity of myxoid tissue is abundant, the lesion will have an elevated signal in the T2-weighted images, whereas if the fibrous component prevails, the mass will appear hypointense. Enhancement of the tumor is correlated to its histology and the areas that do not enhance correspond to necrosis or cystic degeneration.[8] Enhancement is the key difference between myxomas and thrombi; the latter do not enhance except in rare cases of vascularized thrombi.[9] We did not come across such thrombi in our series.

Papillary fibroelastoma is the second most common benign

TABLE 2. Tumor imaging characteristics

	MRI			CT		PET-CT
Lesion	**T1-Weighted**	**T2-Weighted**	**Contrast**	**Pre contrast**	**After contrast**	–
8 Myxomas	Low & Inhomogenous	Inhomogenous	Homogenous Enhancment	35-65 HU	No Enhancement	–
2 Myxomas with abundant myxoid tissue	↓	↑	Homogenous Enhancement	35-65 HU	No Enhancement	–
1 Myxoma totally calcified	↓	↓ in T2-Gradient Echo	Inhomogenous Enhancement	>120 HU	No Enhancement	–
10 Thrombi	Inhomogenous	Inhomogenous	No Enhancement	20-45 HU	No Enhancement	–
1 Lymphoma	Hypointense	Hyperintense	Inhomogenous	–	–	–
1 Melanoma	–	–	–	–	–	↑SUV Value (18) With Homogenous Uptake
1 Paraganglioma	Inhomogenous	↑	Not Performed	–	–	↑SUV Value (15) with Homogenous Uptake
1 Angiosarcoma	–	–	–	70 HU With Hemorrhagic Content	Inhomogenous Enhancement	

HU: Hounsfield Units; SUV: Standard Uptake Value

cardiac tumor. It is made up of fibrous tissue, smooth elastic fiber and muscular cells. The structure is usually small, circular and adherent to the atrioventricular or semilunar valves. There are no definite descriptions on the behavior of the MRI signal and is usually diagnosed by ultrasound. Most are smaller than 1 cm in diameter and attached to valves.[10]

Rhabdomyoma is the most common cardiac tumor seen in childhood. It is considered to be a hamartoma. It may be associated with tuberous sclerosis.[11,12] It accounts for approximately 5% of all primary cardiac lesions. It may present as multiple intramyocardial masses which tend to be pedunculated and are most frequently found in the left ventricle. It tends to regress over time. Conduction problems or obstruction cause the symptoms, rarely hypoxic episodes have also been reported.[13,14] When MRI is performed, it presents the same imaging characteristics as the myocardium on T1-weigheted plain fast spin-echo sequence. It enhances homogeneously, more than the normal myocardium.[1]

Fibromas account for approximately 5% of all primary cardiac lesions and may show progressive growth.[15] They may present as coarse calcifications on CT and reduced signal on T2-weighted sequence in MRI.[16] Lipomas can occur throughout the heart and pericardium. They are frequently round in appearance and vary in size from 2 to 8 cm. Patients may present with a pericardial effusion, supraventricular tachycardia and sudden death. The CT and MRI appearance of fat tissue is characteristic and the tumor characterization is easy.[17]

Primary malignant tumors account for 25-30% of all primary cardiac masses. Sarcomas account for nearly all primary malignant cardiac neoplasms; angiosarcoma is the most common. Patients typically present in their 20-40s with a strong male prevalence. It is associated with a poor prognosis and generally less than a 6-month survival. Our patient survived three weeks post-diagnosis.[18] The right atrium is the predominant site of origin where the mural mass infiltrates the atrial wall and nearly obliterates the chamber. Patients may present with obstructive symptoms, heart failure and/or hemorrhagic pericardial effusion, dyspnea, hemoptysis, arrhythmias and heart block.[19,20] Local invasion of adjacent structures have been described and distant metastases can also occur. CT depicts the tumor as an infiltrating mass extending along the myocardial wall and pericardium. Hemorrhage and necrosis is frequent and responsible for its heterogeneity. MRI detects heterogeneous peripheral contrast enhancement - “sun ray” appearance and nodular areas of increased intensity - “cauliflower appearance”.[21] CT and MRI provide more anatomic

information concerning tumor extension and infiltration than echocardiography.

Rhabdomyosarcoma is the second most frequent primary malignant cardiac tumor and also exhibits a male predominance. Often, multiple lesions involving the valves are seen. On CT it appears as an irregular low attenuation lesion. On MRI it appears as an iso-intense lesion relative to the myocardium on an unenhanced T1-weighted sequence and presents heterogeneous signal intensity after intravenous contrast injection indicating necrosis.[21]

Extremely rare tumors include fibrosarcoma, liposarcoma, and hemangiopericytoma and fibrous histiocytoma. The first one tends to involve multiple sites and valves and has a low attenuation on plain CT. On MRI it is heterogeneous or isointense to myocardium on T1-weighted images. Liposarcoma is a large multilobulated tumor with necrosis. On MRI it is heterogeneous or has high signal intensity presenting with epicardial infiltration.[21]

Primary cardiac lymphoma is a non Hodgkin's lymphoma involving only the heart and pericardium. The commonest presentation is cardiac failure. Manifestations include dyspnea, arrhythmias, superior vena cava obstruction, pericardial effusions, tamponade and chest pain.[22] They commonly arise in the right atrium and ventricle. CT reveals a pericardial effusion and pericardial thickening and usually a polypoid mass or an ill defined infiltrative lesion. On MRI lymphomas have similar or lower intensity than cardiac muscle. Contrast reveals heterogeneous enhancement due to central necrosis.[21] The diagnosis is based on findings of the pericardial fluid and is usually confirmed by biopsy. It is important to have this tumor in the differential diagnosis since it can be treated effectively with chemotherapy. Unfortunately we did not have the opportunity to scan the patient post-treatment.

Metastasis could occur by direct extension, lymphatic or venous hematogenous spread. Direct extension can occur in cases of lung, breast, and esophageal carcinoma. Lymphogenous spread can occur in cases of lung and breast carcinoma. Venous extension can occur in cases of renal cell carcinoma and hepatocellular carcinoma, whereas hematogenous spread is the main route of dissemination in cases of sarcoma, lymphoma, leukemia and melanoma. In our patient with the melanoma metastasis, multiple metastases were identified on the PET/CT exam and did not change the treatment plan. Isolated metastases to the pericardium are rare. Patients may present with pericardial effusion, constriction, and tamponade.[2]

In one of our patients, previously diagnosed by PET-CT with multiple paragangliomas throughout his body, the paraganglioma of the heart was assessed by MRI, but results were inconclusive probably do to the fact that contrast media was not injected (Fig. 7b). PET/CT detected a lesion with high and homogenous 18-FDG uptake in the atrioventricular groove. The uptake was similar to the other body paragangliomas.

"Pseudotumors" usually represent a thrombus. They are the commonest intracardiac masses and may be detected in any cardiac chamber. On enhanced CT, thrombi have a hypodense appearance. On MRI the signal intensity varies according to the age of the thrombus.[1] Intracardiac thrombi can be formed due to changes of surface properties, flow dynamics or changes in blood coagulation. Patients at risk are the ones with prosthetic heart valves, pacemakers, atrial fibrillation or dysfunctional ventricles. Additionally, the presence of a malignant tumor is a significant risk factor for cardiac thrombi, especially in cases of renal cell cancer, extension of tumor thrombus from the kidney through the inferior vena cava and into the right atrium is commonly encountered. On the contrary, hepatocellular carcinoma with tumor thrombus in the inferior vena cava and right atrium is not commonly observed.[23] In one of our cardiac thrombi cases, a right atrial "pseudotumor", first discovered by transthoracic cardiac ultrasound and further investigated by CT, was the primary manifestation of hepatocellular carcinoma. Computed tomography and MRI in this patient revealed a huge primary hepatocellular carcinoma with direct extension of the mass through the inferior vena cava into the right atrium.

Although there is no pediatric experience in our hospital, we will herein briefly review and compare the incidence of cardiac tumors between children and adults. Primary cardiac tumors are rare in the pediatric practice with a prevalence of 0.0017 to 0.28 in autopsy series. In contrast, the incidence of cardiac tumors during fetal life has been reported to be approximately 0.14%.[24] Most of the tumors in childhood are benign, whilst approximately 10% are malignant, with secondary malignant tumors being 10 to 20 times more prevalent than primary ones.[24] The most common cardiac tumor during fetal life and childhood is the rhabdomyoma with an incidence of 60% of all primary cardiac tumors.[24] Teratomas are the second most common tumor in the fetus and neonate after rhabdomyoma (15-19%) and they are most commonly detected in the pericardial cavity, attached to the pulmonary artery and aorta.[24] Fibroma, which is the third most common tumor in children (12-16%) is 5 times more prevalent than in adults, while myxoma is by far less frequent in the pediatric population (2-4% vs 65% in adults).[24] Finally, primary and especially secondary malignant cardiac tumors are rare in both age groups, with rabdomyosarcomα and fibrosarcoma being the most common primary malignant in childhood and sarcomatous in nature tumors in the adults.[24] Concerning secondary malignant tumors, neuroblastoma and melanoma are the most prevalent in children [24] and metastases from lung, breast carcinoma and melanoma are the most prevalent in adults.[25]

CONCLUSION

Based on the findings of the present study and the review

of the literature, we suggest that cardiac CT and MRI can delineate cardiac tumors and provide information essential for adequate diagnosis, staging and treatment planning. Cardiac MRI is a superior tool for cardiac mass differentiation, owing to the detail it provides on soft tissue.[26] In cases of infiltrative tumor, such as angiosarcoma, MRI can provide more details than CT concerning mass extension, due to better soft tissue differentiation. The advantage of multi-slice CT is its speed concerning the time spent by the patient in the gantry and is preferred by the patients, as in the case of the patient with the angiosarcoma who could not complete the exam.

One major *limitation* of our study is that we did not compare our findings with the findings of echocardiography and also we did not use functional techniques with MRI scanning. In general, we find that CT and MRI are better methods for preoperative planning, while cardiac ultrasound is an excellent method for initial diagnosis and follow up of patients.

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